PART III: CONSUMER INFORMATION

**ELAPRASE®**
idursulfase

This leaflet is part III of a three-part "Product Monograph" published when ELAPRASE was approved for sale in Canada and is designed specifically for Consumers. This leaflet is a summary and will not tell you everything about ELAPRASE. Contact your doctor or pharmacist if you have any questions about the drug.

ABOUT THIS MEDICATION

**What the medication is used for:**
ELAPRASE is a long-term enzyme replacement therapy in patients with Hunter syndrome. Treatment with ELAPRASE should be supervised by a physician or other experienced health care provider.

**What it does:**
Patients with Hunter syndrome do not produce enough of their own enzyme, iduronate-2-sulfatase. The reduced iduronate-2-sulfatase levels in patients result in the accumulation of substances called glycosaminoglycans (GAG) in a number of cell types and tissues. ELAPRASE is an enzyme replacement therapy that is intended to restore sufficient levels of enzyme to assist in the removal of these accumulated substances and to reduce further accumulation.

**What the medicinal ingredient is:**
The active substance in ELAPRASE is idursulfase (2 mg/mL). Idursulfase is a form of the human enzyme iduronate-2-sulfatase. It is produced by recombinant DNA technology.

**What the nonmedicinal ingredients are:**
polysorbate 20, sodium chloride, sodium phosphate dibasic heptahydrate, sodium phosphate monobasic monohydrate and water for injection.

**What dosage forms it comes in:**
2 mg/mL concentrate for solution for infusion in a clear, glass vial (bottle).

WARNINGS AND PRECAUTIONS

Serious Warnings and Precautions

- Risk of hypersensitivity reactions: Anaphylactoid/anaphylactic reactions, which have the potential to be life threatening, have been observed in some patients treated with ELAPRASE up to several years after initiating treatment.

- Patients with compromised respiratory function or acute respiratory disease may be at risk of serious exacerbation of their respiratory dysfunction due to infusion related reactions. These patients require additional monitoring. Late-emergent signs and symptoms of anaphylactoid/anaphylactic reactions have been observed after ELAPRASE administration as long as 24 hours after an initial reaction. If an anaphylactoid/anaphylactic reaction occurs, the infusion of ELAPRASE should be immediately suspended and appropriate treatment and observation initiated. The current medical standards for emergency treatment are to be followed. Patients experiencing severe and refractory anaphylactoid reactions may require prolonged observation times.

- Due to the potential for severe infusion reactions appropriate medical support measures should be readily available when ELAPRASE is administered.

If you are treated with ELAPRASE you may experience reactions during or following an infusion. Most infusion reactions are mild or moderate but some may be serious. The most common symptoms are rash, itching, flushing, hypertension, wheezing, cough, headache, abdominal pain, nausea, chest pain, and swelling. Most of the time, you can still be given ELAPRASE even if these symptoms occur. If you experience an allergic side effect following administration of ELAPRASE, you should contact your doctor immediately. You may be given additional medicines such as antihistamines and corticosteroids to treat or help prevent allergic-type reactions.

If severe, allergic-type (hypersensitivity) reactions occur, your doctor may consider stopping the infusion immediately, and should start giving you suitable treatment.

If you have a fever and an acute illness affecting your lungs, your doctor may delay your infusion.

The nature of your genetic mutation may influence your therapeutic response to ELAPRASE, as well as your risk of developing antibodies and infusion-related adverse events; please consult your doctor.

If you are pregnant or breast-feeding, think you may be pregnant or are planning to have a baby, ask your doctor or pharmacist for advice before taking this medicine.
INTERACTIONS WITH THIS MEDICATION

There is no known interaction of ELAPRASE with other medicines.

PROPER USE OF THIS MEDICATION

**Usual dose:**
ELAPRASE has to be diluted in 9 mg/mL (0.9%) sodium chloride solution before use. The usual dose is an infusion of 0.5 mg (half a milligram) for every kg you weigh. This would be about 18 mg or 3 vials (bottles) of ELAPRASE for a 36 kg individual.

After dilution ELAPRASE is given through a vein (drip feed).

The infusion will normally last for 3 hours, which may be gradually reduced to 1 hour if no infusion-related reactions are observed and will be given every week.

**Overdose:**
One patient with Hunter syndrome, who received ELAPRASE at twice the recommended dosage for one and a half years, experienced two anaphylactic reactions over a 3-month period 4.5 years after initiating ELAPRASE treatment.

In case of drug overdose, contact a health care practitioner, hospital emergency department or regional Poison Control Centre immediately, even if there are no symptoms.

SIDE EFFECTS AND WHAT TO DO ABOUT THEM

Like all medicines, ELAPRASE can cause side effects, although not everybody gets them. Most side effects are mild to moderate and generally are associated with the infusion; however some side effects may be serious and may need treatment. Over time the number of these infusion-associated reactions generally decreases.

Very common side effects (more than 1 per 10) are:
- Headache
- Increased blood pressure
- Heartburn
- Chest pain
- Hives, rash, itching
- Fever, and infusion site swelling

Common side effects (more than 1 per 100) are:
- Dizziness, tremor
- Teary eyes
- Changes in the way your heart beats, bluish skin
- Decreased blood pressure, flushing (redness)
- Difficulty breathing, wheezing, blood clot in the lung artery, cough, quickened breathing
- Abdominal pain, nausea, diarrhea, swollen tongue
- Facial swelling, skin lesions (redness, eczema)
- Pain in the joints
- Swelling of the extremities

Allergic reactions have included temporary breathing difficulty, decreased blood pressure, or swelling. In a more severe reaction, in a single patient, a seizure occurred because of a drop in blood oxygen level from difficulty in breathing. Inform your doctor immediately if you have any of these side effects. Patients with compromised respiratory function or acute respiratory disease are at greater risk for infusion-related reactions.

If you notice any side effects not mentioned in this leaflet, please inform your doctor.

A registry (the Hunter Outcome Survey) has been established in order to better understand the variability and progression of the disease and monitoring and evaluation of treatments. All patients are encouraged to participate and advised that their participation may involve long-term follow-up. Information on this registry program is available by calling 1-888-867-7426.

### SERIOUS SIDE EFFECTS, HOW OFTEN THEY HAPPEN AND WHAT TO DO ABOUT THEM

<table>
<thead>
<tr>
<th>Symptom / effect</th>
<th>Talk with your doctor or pharmacist</th>
<th>Stop taking drug and call your doctor or pharmacist</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Common/Very common (occurring in ≥5% in controlled clinical studies)</strong></td>
<td></td>
<td></td>
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<tr>
<td>Fever</td>
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<td>Difficulty breathing</td>
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<td>Bluish skin discoloration</td>
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<tr>
<td>Rash or Hives</td>
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<tr>
<td>Seizure, blood clot in the lungs</td>
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<tr>
<td>Missed or extra heartbeats</td>
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This is not a complete list of side effects. For any unexpected effects while taking ELAPRASE, contact your doctor or pharmacist.

HOW TO STORE IT

Store at 2°C to 8°C (in a refrigerator), and protect from light.

Do not freeze or shake.
Reporting Side effects
You can help improve the safe use of health products for Canadians by reporting serious and unexpected side effects to Health Canada. Your report may help to identify new side effects and change the product safety information.

3 ways to report:
- Online at www.santecanada.gc.ca/medeffet
- By calling 1-866-234-2345 (toll-free);
- By completing a Patient Side Effect Reporting Form and sending it by:
  - Fax to 1-866-678-6789 (toll-free), or
  - Mail to: Canada Vigilance Program
    Health Canada, Postal Locator 0701E
    Ottawa, ON
    K1A 0K9

Postage paid labels and the Patient Side Effects Reporting Form are available at www.healthcanada.gc.ca/medeffect

Note: Contact your health professional if you need information about how to manage your side effects The Canada Vigilance Program does not provide medical advice.

MORE INFORMATION

If you want more information about ELAPRASE:
- Talk to your healthcare professional
- Find the full product monograph that is prepared for healthcare professionals and includes this Consumer Information by visiting the Health Canada website http://hc-gc.ca, the manufacturer’s website www.shirecanada.com, or by calling 1-888-867-7426

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